DIATAXIA CEREBRALIS INFANTILIS: THE ATAXIC TYPE OF CEREBRAL BIRTH PALSY.*

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The classification of the cerebral palsies of early life has engaged the attention of neurologists and psychiatrists for a number of years. A great many clinical types have been described, and in recent years there has been considerable progress in our knowledge of their various causes and underlying pathological changes. The subject, however, still presents many obscurities in etiology, localization and symptomatology.

The cases have been variously grouped in accordance with the etiology, the nature of the pathological changes or the clinical symptoms presented. For all practical purposes the clinical classification is the important one and the most useful, as postmortem examination and histological study are usually necessary to determine the true pathology of any case and the original causative factors are often obscure and difficult to determine. This is especially true of the congenital cerebral defects and malformations resulting from disease of the fetus during the period of intra-uterine life.

The common clinical type of infantile cerebral palsy is the spastic form. This may appear as hemiplegia, paraplegia or triplegia, and very frequently as a cerebral diplegia. It is characterized by paralysis with various spastic phenomena, either alone or in association with other cerebral symptoms, such as choreiform manifestations, ataxia, tremor, athetosis, mental defect, epilepsy and disturbance of the special senses.

A classical description of this large clinical group and its relationship to injuries received at birth was given by an English physician, Dr. Little, in 1862. In recognition of this important contribution, those cases of *cerebral spastic paralysis* resulting from injury to the brain at birth are very commonly known under the generic title of Little's disease. Little's conception that certain abnormal types of parturition, especially those associated with difficult and prolonged labor, produced hemorrhages over the surface of the brain which interfered with the normal functions and development of motility has been upheld by many subsequent investigators. Prominent among these may be mentioned the names of Sarah McNutt, Freud, Gowers, Railton and Kundrat.

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Little also pointed out the importance of premature birth as a cause of cerebral spastic paralysis, and some observers, notably Brissaud, would limit the term Little's disease to this comparatively small group of cases, in which the development of the pyramidal tracts has been checked or retarded by premature delivery. This limitation of Little's conception in the opinion of many writers is not justified, and a perusal of his original monograph will, I believe, convince most readers that the essential idea underlying his work was the relationship of all forms of abnormal birth to the cerebral spastic palsies of early life.

Förster, in 1909, described in some detail an unusual clinical type of cerebral diplegia which he termed the *atonic-astasic type* of infantile cerebral palsy. This group stands in striking contrast to the spastic type of Little, and is characterized by motor paralysis with flaccidity. There is inability to stand or walk (astasia-abasia), great difficulty in articulation, not infrequently mutism, and a marked mental defect. A certain degree of individual movement of the extremities is possible, but all the larger and more complicated motor activities are impossible, owing to a defective action of antagonistic and synergic groups of muscles. Slight spastic phenomena are sometimes noted in the lower extremities, but the essential quality of the paralysis is flaccid and atonic.

Many of the symptoms in this group of cases suggest a cerebellar origin, and Förster was at first inclined to attribute this origin to the symptomatology until later two of his cases subjected to postmortem examination revealed an extensive lobar sclerosis of the frontal region, the cerebellum appearing quite normal on macroscopic examination. Unfortunately, no microscopic studies were recorded.

A few years later, in 1913, Pierce Clark reported similar cases under the rather comprehensive title, "Cerebrocerebellar Diplegia of Flaccid Atonic Type," and concluded on purely clinical grounds that the lesion was probably a combined one, involving both the cerebrum and the cerebellum, notwithstanding the negative cerebellar findings in Förster's cases.

Clark assumed that a cerebellar lesion alone would be amply compensated for by the cerebrum if the cerebral structures were normal, and therefore a combination of cerebral and cerebellar symptoms as occurs in this group of cases would be in favor of a combined lesion of these two structures. Against this point of view, however, may be mentioned the normal appearance of the cerebellum in Förster's cases and also the fact that grave disturbances of muscle tone and coördination may result from lesions of the cerebral cortex. Accordingly, the term cerebrocerebellar diplegia in the present state of our knowledge would seem hardly justified and the simple clinical designation atonic or flaccid type of cerebral diplegia as used by Förster is, I believe, preferable.

Batten and von Wyss have also recorded examples of this flaccid type, and there can be no question as to its distinctive clinical characteristics and marked contrast to the spastic types of Little.

Under the title "Congenital Cerebellar Ataxia," Dr. Batten has described what he terms a pure *cerebellar type* of diplegia. This group is characterized by ataxia of the cerebellar type, which displays, however, a decided tendency toward recovery. There is little or no mental defect, and the clinical picture is one of pure ataxia of cerebellar origin involving speech, the gait and station and various movements of the extremities.

Therefore, according to our present classification, the cerebral diplegias of childhood may assume one or other of the following clinical forms; the *spastic* type of Little, the *atonic* or *flaccid* type of Förster and a *cerebellar* type as described by Batten. These may exist as such in pure form or may occur in various combinations. They may also present a variety of complications and associated symptoms such as epilepsy, mental defect, choreiform and athetoid manifestations, which give to this whole chapter a large and varied symptomatology.

The group of the cerebral diplegias to which I would direct attention as a distinct clinical variety is an *ataxic type of cerebral birth palsy*. The ataxia is bilateral in distribution and is characterized by a generalized disturbance of coördination involving the gait and station, the use of the arms and legs, and the muscles subserving the function of speech. The incoördination persists in the recumbent posture and has the characteristics of a true ataxia. It is, I believe, of cortical origin. There is neither paralysis nor spasticity, and the cases observed were free from serious mental defect or epilepsy. In the absence of any direct pathological evidence I would suggest, as the probable etiological factor, an injury to the fetal head received at birth, producing rupture or thrombosis of the cerebral veins, with limitation of the lesions to the cortex of the parietal lobes.

The clinical picture is one of pure *cerebral diataxia*, and may be regarded as the sensory equivalent of *cerebral diplegia*. In my opinion, however, the vascular lesion, instead of being limited to the motor area as in spastic diplegia, is situated more posteriorly behind the fissure of Rolando in the sensory sphere of the cortex, thus implicating the centers and commissural systems which are engaged in the reception and transmission of the memories of movement.

The parietal lobe is the great receptive center for the registration of sensory stimuli, representing movements and the various combinations of movement, and a bilateral lesion in this area would necessarily impair in greater or lesser degree the normal development of motility and coördination.

Case I.—The patient, E. C., was a boy, aged twelve years, born of healthy parents. There was no history of any familial nervous or mental disease. With the exception of measles, chickenpox and tonsillitis there was no history of any serious illness.

He was an only child, born at term, the labor being unusually difficult and prolonged. The mother stated that she was thirty-five years old at the time and was in labor for forty-eight hours. The membranes had ruptured early and delivery was eventually effected by forceps. The child was born blue and asphyxiated and did not cry for several days. There were general convulsive seizures during the first few days, but none subsequently. The head was said to have been out of shape and deformed, but gradually assumed a normal symmetry. An effort was made to feed the child by the breast, which was unsuccessful and had to be abandoned.

Very early in the child's life it was noted that the movements of the extremities and trunk were weak and awkward. Swallowing was performed without apparent difficulty; there was, however, a tendency to drooling, which persisted for several years. It was noted that the child did not sit up or show much tendency to creep until the end of the second year. The movements then were awkward, unsteady and accompanied by unusual efforts. Unsteadiness of the movements was noted quite early and has persisted up to the present time. Both sides were involved, but the right arm seemed to be more affected than the left, and the legs showed a greater degree of disability than the arms.

The child did not speak until the third year, and then only uttered inarticulate sounds. He has shown slight but steady improvement from year to year in speech, in his ability to sit or stand and in the use of the arms. He could not stand alone until the sixth year, and is still unable to walk alone.

The general health and intelligence are both good. The mother insists that the child is bright and learns readily. He is quite cleanly in his habits, and is able to control the sphincters. He is not able to feed himself or dress himself on account of the severe motor ataxia. He is good tempered, and is interested in the books and games suitable to his age. He has had no convulsions since the initial spasms after the birth, which lasted several days. The special senses are apparently not affected.

Examination, May, 1916. The boy is of good size, well nourished, and appears in excellent general health. The muscles are moderately well developed. There is a slight tendency to motor restlessness and atactiform movements unless supported and in a relaxed position. This unrest is increased by any action, however slight, and by mental excitement. There is no true chorea or athetosis, but the ataxic disturbance has a somewhat choreiform character when the motor agitation is extreme.

He is able to stand by holding on to a chair, but not unsupported. While standing in this position there are swaying, unsteady movements of the trunk and arms in the effort to maintain the equilibrium. The head also shows ataxic oscillations. On attempting to take a few steps, even with assistance, the ataxic disorder becomes extreme and he falls backward or to the side and has to be supported; occasionally the legs give way under him.

In testing the movements of the upper extremities a very marked incoördination is demonstrable. On attempting to carry the finger to the tip of the nose there are irregular atactiform movements of the extremity and compensatory or associated movements in other parts of the body—for example, the head and trunk. This disorder of motility does not resemble an intention tremor but is a gross disturbance of the power of coördination. It is especially well marked if an effort is made to grasp a small object or to draw a line with a pencil. Such movements are carried out in a brusque and awkward manner, and any breakable object would surely be sacrificed if he were allowed to reach for it. If the attempt is made to hold out the arms with the hands and fingers extended there are irregular atactiform movements and changes in the relative position of the extremities, which are somewhat increased by closure of the eyes.

The alternating rhythmical movements of the wrists or fingers, as a result of the ataxic disturbance, are impaired. Dysdiadokokinesis is therefore present.

In the recumbent posture the lower extremities show a similar atactiform disturbance. If the attempt is made to place the heel upon the opposite knee there is a marked incoördination and uncertainty of movement, as might be observed in an advanced case of spinal ataxia.

Similarly, if the legs and arms are elevated while lying flat on the back and the effort is made to fix them in this position the extremities are moved about in extreme incoördination, and frequently the balance is lost and he rolls over to one side. The right arm and leg showed a somewhat greater degree of involvement than the left, which coincided with the early history and observations of the mother.

There is no demonstrable weakness or paralysis of the limbs. Indeed, the gross motor power of the lower and upper extremities is well preserved, except as modified by the ataxia. There is no atrophy. There is not the slightest evidence of spasticity in either the upper or lower extremities. Instead, the tonus of the musculature is moderately diminished.

The knee-jerks are present, equal and not exaggerated. The Achilles-jerks are inactive and only elicited with difficulty. The supinator jerks are not elicitable. The biceps- and triceps-jerks are inactive and variable. There are no periosteal reflexes in either the upper or lower extremities and no clonus or spastic phenomena. The abdominal, cremasteric and plantar reflexes are present and of normal intensity. The Babinski phenomenon is absent.

The sensation, both superficial and deep, is apparently undisturbed. Touch, pain and temperature are recognized promptly, and so far as could be determined were well localized. The deep sensibility also appears normal. Pressure of the muscle induces pain and the sense of position is preserved. He recognizes the direction of passive movements of the toes, fingers and of the larger joints, promptly and without error. The sense of vibration is acute.

The pupils are equal and react promptly to light and accommodation. The pupillary skin reflex is active.

There is no nystagmus and the optic nerves are normal. On attempting to fix an object with the eyes there are slight rolling movements of the eyeballs, apparently of an ataxic nature, and associated atactiform oscillations of the head. There is no paralysis of the cranial nerves. The tongue is protruded in the median line, the palate is promptly innervated and the movements of the jaw are normal in power and extent. Facial innervation is equal on the two sides.

There is a considerable degree of dysarthria, and it is rather difficult to understand the spoken words. The mother, however, interprets perfectly and can carry on a very intelligent conversation with the boy. He can name ordinary objects placed in the hands, and there is no astereognosis. Sight, hearing and smell are normal.

The heart and lungs are normal.

The urine is free from albumin and sugar. The Wassermann test of the blood is negative.

NOTE.—The above findings were confirmed at several subsequent examinations. The essential disturbance was one of incoördination without paralysis or evidences of spasticity, which affected nearly all voluntary movements. While in the recumbent posture, with the musculature relaxed, there were no evidences of chorea or athetosis. On making the effort to stand or walk, and in certain postures, there were incoördinate movements which suggested a choreiform tendency, which, however, appeared to be dependent upon the degree of the ataxic disturbance. The right side showed a greater degree of involvement than the left. The tonus of the muscles was moderately reduced. There was, however, no undue flexibility of the extremities as occurs in the higher grades of hypotonicity. The knee-jerks were readily elicitable and not exaggerated. The Achilles-jerks could usually be elicited, but with difficulty, the same being true of the reflexes of the upper extremities. The plantar reflex was always of the normal type, and a true Babinski reflex was not obtained.

So far as could be determined the general sensations, both superficial and deep, were well preserved.

Case II.—The patient, L. H., was a girl, aged six and a half years. The family history was good and there was no tendency to hereditary or familial diseases. The parents were healthy and were not related. The mother had given birth to two other children without special difficulty, and there had been no miscarriages.

The patient was the third child and the pregnancy had progressed without untoward complications. The birth occurred at term and the labor was extremely slow and tedious. After twelve to fourteen hours a faulty presentation was discovered and the delivery was effected by means of forceps. The child was born asphyxiated, and it was feared at the time would not survive. Artificial respiration was necessary and the child did not cry or take the breast for about two weeks. After this she took the breast, but only with some difficulty, and the cry was weak and feeble. The subsequent development was slow and retarded. The child could not sit up until two years of age, and then was unsteady and swaying, often falling backward or to one side. About this time she was able to creep, but with difficulty. Movements of the arms and legs were awkward and unsteady. Attempts to stand were not successful until the third year, and then only by holding on with the hands and accompanied by swaying of the body and frequent falls. Attempts at walking were only possible in the latter part of the third year, and then required careful assistance, as falls were frequent and often severe.

At the age of three years the child could speak a few words, but indistinctly, and only the mother could understand them. Swallowing occurred without difficulty and the special senses were apparently normal. The child was bright and took an interest in her surroundings, but was backward as compared with other children at the same age. On attempting to grasp an object with the hand the movements were uncertain and awkward.

The child had shown decided but gradual improvement. She had acquired better control of her movements and articulation was more distinct. She was fairly intelligent for her age but still somewhat backward in many things, which the mother attributed to her physical disabilities. There was no history of convulsive seizures.

Physical Examination October, 1914. The child is well nourished and fairly well developed for her age. While sitting quietly in a chair the extremities and trunk are motionless. There is a slight tendency to swaying and oscillation of the head, especially with any movement, such as fixation of an object with the eyes or when speaking.

Any attempt to raise an arm or leg evokes awkward ataxic movements of an incoördinate character. On attempting to grasp an object with the hands there is a series of irregular, incoördinate movements; the fingers are overextended, the hand is thrown about in an awkward manner and the object is grasped brusquely. There is no weakness of the movement or of the grasp—it is a pure ataxia of movement or incoördination. The disturbance is about equal on the two sides.

The child understands spoken language and talks, but articulation is dysarthric and indistinct and accompanied by slight atactiform movements of the head and trunk, especially if standing. In standing there is manifested a distinct disturbance of equilibrium. She stands on a broad base, with swaying of the trunk and various compensatory movements of the arms. This static ataxia is increased slightly by closure of the eyes. On attempting to walk a definite disturbance of the gait immediately becomes apparent. There is unsteadiness of equilibrium, pitching to and fro and great uncertainty of movement. In appearance it suggests very strongly the titubating gait of cerebellar disease. The legs are held far apart, the head erect and the arms used to balance. The movements of the legs are awkward and ataxic. On attempting to stand quietly there is a slight choreic unrest, which, however, is ataxic in origin and is not noticeable in a quiet and relaxed position. The ataxia does not disappear in the recumbent position. For example, on attempting the finger-nose test or the heel-knee test, while flat on the back, the incoördination of movements is scarcely less marked than in the erect posture or sitting in a chair. There is no cessation or diminution of the ataxia when the recumbent posture is assumed. In this position, if the arms and legs are elevated, and the attempt made to hold all four extremities upright, with the eyes closed, incoördinate, ataxic movements persist with unabated vigor. If the hands and arms are held outstretched, while sitting in a chair there are ataxic movements of the upper extremities and some swaying movements of the trunk. Rhythmic alternating movements of the wrist and fingers are performed slowly and awkwardly. Evidences of dysdiadokokinesis are therefore present.

The cranial nerves are normal. The tongue is protruded in the median line, the facial innervation, movements of the soft palate and of the mandible are all normal. These movements are associated with slight atactiform manifestations of the head and trunk.

The optic nerves are normal. There is no nystagmus and the pupils are equal and react to light.

There is no paralysis and all movements show a normal degree of power. The muscles are well developed and show no atrophy.

There is no spasticity in either the arms or legs and no clonus. The tonus of the muscles is slightly reduced but the hypotonicity is of moderate degree. There is plantar flexion of the toes; the Babinski reflex is not present.

The knee-jerks are present, equal and of normal intensity. The Achilles-jerks are faint and at times not elicitable with certainty. The arm reflexes, supinator, biceps- and triceps-jerks, are inactive and not definitely elicitable. The abdominal reflexes are present.

General sensation was apparently normal. Touch, pain and temperature stimuli were correctly interpreted and, so far as could be determined, were well localized.

The deep sensibility and sense of posture were also preserved.

The child could recognize the movements of the toes whether up or down, but if the order was given to place the left limb in the same position as the right, either the arm or leg, the attempt was made and sustained awkwardly and in an uncertain manner, probably on account of the ataxia. Sight, hearing and smell were not affected. There was no astereognosis. Such common objects as a coin, penknife or a key were promptly recognized in either hand.

There was no special mental defect; the child was good natured, responsive and well behaved and was not far below the mental average for her age.

Heart and lungs were normal. Wassermann test of the blood was negative. Urine negative.

NOTE.—This case, like the preceding, presented symptoms of a pure ataxic disturbance which affected nearly all voluntary movements, such as standing, walking and the individual activities of the extremities. The gait was very suggestive of a cerebellar ataxia but both the upper and lower extremities showed a definite ataxia which persisted in the recumbent posture. Lying flat on the back, with no demands upon the function of equilibrium, it was impossible to hold the extremities quietly in an elevated position as in cases of pure cerebellar disease, the ataxic disturbance persisting as in a case of spinal ataxia. There was no demonstrable weakness of the extremities and no spasticity. The sensibility was normal. There was no true chorea, athetosis or intention tremor. The slight choreiform tendency did not suggest true chorea but resembled rather the choreic unrest which is sometimes present in cases of advanced Friedreich's ataxia. The disorder of motility was an inability to regulate and coördinate voluntary movement. The condition was manifested soon after birth and was characterized by steady improvement. There was therefore no tendency to progression or aggravation of symptoms.

CASE III.—The patient, A. H., a girl, aged eleven years, was the last of seven children. The circumstances attending the birth were abnormal, the membranes ruptured quite early and labor was unusually prolonged and severe. The child was eventually delivered with forceps in an asphyxiated condition and was resuscitated with difficulty. She was said to have been "blue" for several hours after birth and the imprint of the forceps remained over the temporal region for a week or more. She did not take the breast readily and was nourished by the bottle. During the early months of life she was restless and difficult to quiet, crying a great deal, and was physically frail.

It was soon observed that the motor development was not normal. Such acts as holding up the head, sitting and attempts to crawl and walk were all considerably delayed, as was also speech.

She was unable to stand before the sixth year and then only with assistance. The movements of the arms were noted to be irregular and unsteady early in the second year and articulation was indistinct up to the fourth year, but has shown a steady improvement since that time.

With the exception of summer diarrhea and chicken-pox the child has had no illnesses of note. The other children of the family were in good health. The father died of pneumonia and no history of any neuropathic heredity could be elicited.

The mother states that while somewhat backward for her age the child is bright, has learned to read a little and is especially fond of music. The special senses were not affected. There has been a gradual improvement in the motor disability from year to year, especially in speaking and walking. There has been no history of any convulsive seizures, excepting immediately after the birth, when there was an indefinite history of several spasmodic attacks.

Examination, November, 1912. The patient is thin and somewhat pallid; the teeth are uneven and defective.

The skeletal development is rather slight and fragile. The muscles are well formed and, although small, are not atrophic. On attempting to rise from a sitting posture there is a very evident disturbance of gait and station. She stands on a broad base, the trunk swaying from side to side and balancing with the arms. After a few moments she becomes somewhat adjusted to the change of posture, but slight atactiform and balancing movements persist. Closure of the eyes increases somewhat the degree of static ataxia.

While walking there is apparent a marked disorder of movement. The legs are lifted and thrown out in an awkward, ataxic manner and there are numerous compensatory lurches and balancing movements of the head, trunk and arms. She is, however, able to walk alone and with a fair degree of security, and does not fall. While sitting in a relaxed posture in a chair the musculature is quiet, and only occasionally slight atactiform movements of the head or arms are observed. On attempting to stand or walk or on initiating any movement of the extremities a considerable degree of ataxia results which is somewhat choreiform in appearance, but occurs only as an accompaniment of movement.

There is a general condition of marked incoördination. On attempting to grasp an object with the hand or on placing a finger on the tip of the nose a considerable degree of ataxia of the arms is apparent, which is increased by the associated atactiform movements in other regions of the body, as, for example, the head and trunk. The two arms are about equally affected.

There is also marked retardation and awkwardness in performing alternating rhythmic movements of the wrist and fingers (dysdiadokokinesis). There is no weakness of the upper extremities and the gross motor power is apparently normal.

The supinator reflexes are present and not exaggerated. The biceps- and triceps-jerks are not elicitable. The muscles are somewhat relaxed and free from any spastic phenomena.

On attempting to hold the outstretched arms and hands at rest there occur slight ataxic movements which are increased by closure of the eyes. Objects placed in either hand with the eyes closed are promptly recognized and the stereognostic sense is unimpaired. The direction of passive movements of the fingers, wrist and elbow are promptly recognized with the eyes closed. With the eyes closed an attempt to place the segments of the upper extremity on one side in the same position as the other cannot be accomplished, owing to the ataxia.

The gross motor power of the lower extremities is well preserved and there is no special atrophy of the musculature. Passive movements are quite free, the muscles are relaxed and there are no evidences of spasticity. There is, however, marked incoördination of movement in the recumbent posture. The attempt to place the heel upon the knee or to elevate the extremity in a straight line produces a marked irregularity and incoördination of movement, which is sometimes accompanied by slight atactiform jerking movements of the head, trunk or upper extremities. The knee-jerks are elicitable and about equal on the two sides. They are not exaggerated and the responses are rather subnormal. The Achilles-jerks are faint and not always elicitable. The plantar reflexes are of the normal type (no Babinski).

Lying perfectly quiet and in a relaxed posture no choreiform or athetoid movements are present. If the arms and legs are then simultaneously elevated and the attempt is made to hold them in this position a great disorder of coördination immediately results usually with loss of balance and a tendency to roll over. The abdominal and epigastric reflexes are present and active on both sides.

The motor cranial nerves are normal. There is no paresis of the ocular, facial or palatal musculature. On protrusion of the tongue there are slight atactiform movements of the head and trunk, and in a lesser degree on testing the facial innervation or on fixing an object with the eyes.

There is no nystagmus. The pupils are equal and react to light and accommodation. The corneal and conjunctival reflexes are active. The jaw-jerk is not elicitable. Articulation is thick and indistinct. This is an ataxic, dysarthric disturbance of speech, and to the untrained ear is very difficult to understand.

There is no forced laughter, but the act of laughing is accompanied by ataxic movements of the head and trunk and to some degree of the extremities. The acts of deglutition and mastication are also accompanied by slight ataxic disturbance.

The special senses are not affected. Vision, hearing, and the sense of smell are normal. The optic nerves are normal.

The general sensibility is apparently undisturbed. Tactile, pain and temperature sensations are normal, and their localization, so far as could be determined, was fairly accurate. The posture sense of the upper and lower extremities was apparently preserved and upward and downward movements of the joints were promptly recognized.

Heart and lungs are normal. Urine negative.

Mentally the child was bright and responsive and carried out the various tests promptly and intelligently. She was able to spell and could read short sentences, and the mother stated that she was making steady progress. She was cleanly in all of her habits and has had good control of the sphincters since early childhood. There has been a marked improvement in the child's condition during the past few years, but she still required constant care and supervision because of the general motor disability.

Resumé of the Clinical Reports. The cases which have just been described are very similar in their etiology and general clinical characteristics and constitute, I believe, a well-defined clinical type of cerebral birth palsy. They represent an ataxic type of cerebral diplegia which stands in striking contrast to the spastic type of Little and the flaccid or atonic type of Förster.

In all of the cases there was a history of severe injury at birth followed by retarded and defective development of the power of motor coördination. There was no paralysis, and if such existed after birth it was of a transient nature and the gross motor power was eventually completely restored. There were no evidences of spasticity. The active and passive movements as well as the reflexes revealed none of the characteristic signs of pyramidal tract involvement. The tonus of the muscles was slightly reduced and the tendon reflexes showed a corresponding reaction, and were moderately diminished and difficult to elicit. There was no undue flexibility of the extremities as occurs in the severer forms of hypotonia.

The essential symptom was a generalized ataxic disturbance affecting the gait, station, articulation and the use of the individual members. This was present in all positions, including the recumbent posture, and I would particularly emphasize the persistence of an ataxic disturbance of the arms and legs in the recumbent posture when no effort was required to maintain the equilibrium. The ataxic disturbance was also increased by closure of the eyes, by mental excitement and by any effort to carry out precise and definite movements.

The disturbance of coördination was bilateral and fairly symmetrical, but one side may show a greater degree of involvement. It affected the finer movements of the hands and fingers as well as movements of the larger joints, and seemed to be equally distributed in this respect. There was, at times, a slight tendency to motor unrest, somewhat suggestive of a choreic disturbance, which I regard as a manifestation of ataxia and the effort to maintain posture. The disorder of motility was clearly ataxic, and was characterized by an inability to carry out precise and definite coördinate movements. It was not in any sense a true chorea or athetosis, the restless atactiform movements in certain postures merely suggesting the choreiform tendency. Dysarthria was also a well-marked symptom, and was regarded as part of the ataxia. There was no nystagmus. The sensibility, both superficial and deep, so far as could be determined by the usual clinical tests, was quite normal. Especially noteworthy was the preservation of the sense of posture and the stereognostic sense. In none of the cases was there a serious mental defect. The retardation was no more than might follow a severe disturbance of speech and incoördination and the restriction of opportunity imposed upon the child.

In none of the cases was there any tendency to grand mal or petit mal seizures. The convulsions occurring immediately after the birth had manifested no tendency to recur in later life.

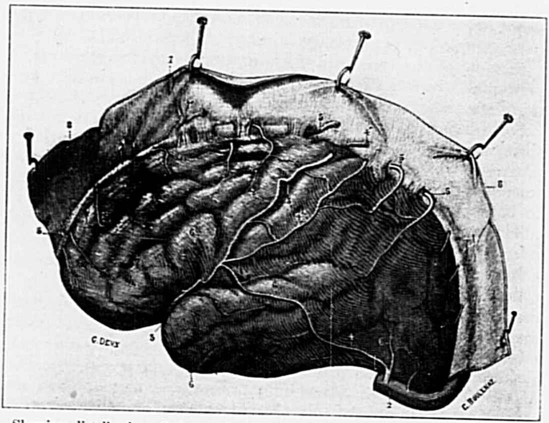
Some improvement had taken place in all of the patients, and there were no indications of an increasing mental deterioration or progression of other symptoms. In this respect the ataxic form resembles the spastic type of Little with its well-recognized tendency to improvement.

This very striking disturbance of motor incoördination I would refer to a disorder of the cortical functions, probably the result of a bilateral injury to the parietal area at birth. A lesion of this character would interfere with the development of the higher cortical centers engaged in the reception and elaboration of the memories of movements, and produce a *pure cortical ataxia;* or it might interrupt the commissural symptoms by which these memories are communicated to the motor area for the proper regulation of coördination, with a resulting *transcortical ataxia.* In either event a serious loss of the control and regulation of movement would of necessity result.

The primordial cortical centers which receive muscle and posture sensibility are not affected, for the general sensibility, both superficial and deep, is apparently preserved. It is probable therefore that the cortical centers subserving the function of common sensibility are either not included within the limits of the destructive lesion or, if they are injured, their function is subsequently restored by the compensatory activities of other centers, cortical and infracortical.

Pathological Considerations. The pathology of this group of cases in the absence of definite pathological evidence may only be surmised. I believe, however, that we are justified in the assumption that the lesions are of the same general character as have been found associated with other types of the cerebral birth palsies, differing only in their localization.

The pathological studies of Kundrat have shown that thrombosis of the superior longitudinal sinus with occlusion or rupture of the cerebral veins is a by no means uncommon sequel of injury to the child's head during birth. This form of lesion is very probably favored, if not directly induced, by overlapping of the parietal bones during the process of molding of the fetal head. During the course of a prolonged and difficult labor, instrumental delivery, or precipitate parturition, the delicate cerebral veins may be torn from their connections as they traverse the intermeningeal space to enter the sinus, with consecutive hemorrhage over the surface of the brain, either subdural or intermeningeal.



Showing distribution of the cerebral veins on the external surface of the hemisphere. The dura mater is lifted to show the termination of the veins in the superior longitudinal sinus. *1*, superior longitudinal sinus; *2*, horizontal portion of lateral sinus; *3*, large anastomotic vein of Trolard; *4*, anastomotic vein of Labbé; 4^1, anastomosis between the vein of Trolard and the superior longitudinal sinus; *5*, *5*, *5*, ascending veins of the hemisphere; *6*, *6*, *6*, descending veins; *7*, branch of the middle meningeal anastomosing at 7^1 with an ascending cerebral vein as it enters the sinus; *8*, dura mater. (From Testut's Anatomy.)

The subdural type of hemorrhage is usually massive, consisting of a single large clot, and is commonly unilateral. Intermeningeal hemorrhages are more diffuse, often bilateral, and consist of a thin film-like clot of blood confined by the piarachnoid. This is greatest in the region of the superior longitudinal fissure where the cerebral veins cross to the sinus and gradually tapers off as it passes over the surface of the hemisphere toward the fissure of Sylvius. As the larger veins run their course in relation to the fissures there is a natural tendency for the blood to follow and gravitate in the depths of the larger cerebral sulci. Limitation of the vascular lesion to the region of the fissure of Rolando would explain the occurrence of spastic diplegia, and a similar explanation might be offered for the manifestations of cerebral diataxia, namely, thrombosis or hemorrhage limited to the parietal veins of the cerebral cortex (see figure).

Kundrat also emphasized the importance of structural peculiarities of the margins of the parietal bones and intervening membrane to injuries of the sinus. He expressed the belief that thrombosis and hemorrhages of venous origin are much more likely to occur in infants with firm bony edges and narrow interosseous membranes, because of the mechanical interference with overlap in the process of molding of the fetal head. A narrow interstitial membrane and parietal bones, with thick heavy margins, would render the sinus liable to the dangers of compression, with consecutive thrombosis or hemorrhage of the cortical veins. Kundrat also performed the interesting experiment of attempting an injection of the cerebral veins through the exposed sinus while holding the parietal bones firmly in a position of overlap, and found that the backward flow of blood through the sinus was effectively blocked by the compressive action of the bones. The dangers of such a contingency in long-protracted labor are obvious notwithstanding the rich collateral circulation of the cerebral venous system.

The essential lesions of the cerebral birth palsy consist therefore of hemorrhage either subdural or intermeningeal, with more or less thrombotic softening of the cerebral cortex from obstruction to the cerebral venous system. These may occur in various combinations. In grave cases lacerations of the brain substance, ventricular and intracerebral apoplexy and hemorrhages at the base of the brain are occasionally observed. Involvement of the cortical venous system is, however, the most important from the clinical point of view, as the severer forms of injury are usually rapidly fatal and therefore have little more than a pathological interest.

Studies of the cerebral cortex in cases of sinus thrombosis also show that intermeningeal hemorrhage is by no means the most important factor in producing the serious consequences of birth palsy, but that grave and permanent changes in the cortex itself may result. Thrombosis of the cerebral veins produces retrograde engorgement of blood, with punctate hemorrhages in the corresponding vascular distribution, and is associated with edema, perivascular engorgement and degeneration of the ganglion cells. (Hunt.) Although a certain degree of collateral circulation may be established, the delicate cells and neurons of the superficial layers of the cerebral cortex would inevitably suffer considerable damage.

Any limitation of the venous thrombosis to a definite vascular distribution would play a very important role in the causation of special clinical types and would explain the occurrence of purely spastic or purely ataxic types of this affection.

In the majority of cases the effusion or thrombosis takes place in the motor areas, producing the spastic type described by Little. That this lesion may be quite circumscribed is shown by the not infrequent occurrence of purely spastic states without mental defect, epilepsy, chorea or athetosis.

The ataxic type of cerebral birth palsy is, I believe, the result of a more or less circumscribed lesion of similar nature within the confines of the parietal area behind the fissure of Rolando. The anatomical distribution of the parietal veins of the cerebral cortex render such a lesion not improbable, the effusion of blood and cortical degenerations interfering with the proper development of the various centers and commissural systems which subserve the higher functions of the coördination of movement (see figures). A lesion of this character might affect a dissociation of the sensory and motor spheres of the cortex, producing a condition in which motion would be possible and the sensibility preserved, and yet the harmony and coördination of movement would be lost, a form of *transcortical ataxia* in which there is an inability to convey to the motor area the complicated memories of movements and their combinations, so essential for a precise coördinate act.

That the parietal lobes contain important centers for muscle sensibility and the memories of movements has been shown by a number of observers and is accepted by all authorities on cerebral localization. The parietal sensory area includes the postcentral convolution, the superior parietal lobule (PI) and the inferior parietal lobule (PII) and portions of the supramarginal and angular gyri. It includes as well the precuneus on the mesial surface of the hemisphere.

In this region are stored not only the sensations of individual joints and muscles, the proprioceptive sensibility proper, but also the higher and more complicated motor memories, the various acquired psychic memories of movements which are essential for the development of a finer and more delicate system of coördination.

Von Monokow has suggested that the *fasciculus centroparietalis*, an association bundle uniting the angular gyrus and the Rolandic area, may play a role in the cortical association of muscle sensibility. He regards the parietal convolutions PI and PII and their association tracts as of special importance for the representation of movements and combinations of movements. The sensations of the individual muscle groups and segments and tactile sensibility he would localize in the postcentral convolution.

It is now generally held that the simple perception of deep sensibility, including the muscle sense and the sense of posture, are localized in the ascending parietal convolutions (postcentral gyrus), probably in segmental relation to the corresponding motor centers of the central convolutions (Mills), while the higher psychic perceptions of motor activity, namely, the memories of movements and combinations of movements are situated more posteriorly in the superior and inferior parietal lobule, a region which is concerned with the registration of higher motor memories.

Indeed, it is quite conceivable that the centers which are engaged in the perception of deep sensibility might escape or recover from the effects of a lesion, and yet the higher centers and commissural systems which register and transmit the more complicated motor memories be seriously impaired.

The view which has just been expressed as to the pathology and localization of the lesions in cases of cerebral birth palsy receives a certain confirmation from the clinical and pathological investigations of Holmes and Sargent of injuries to the superior longitudinal sinus from gunshot wounds of the head. In their large series of cases, comprising seventy observations, the conspicuous feature of the clinical picture was a bilateral spastic paralysis, with marked rigidity involving the legs, trunk and arms in the order named. Articulation and the movements of the face were usually not affected. The paralysis was greatest in the legs and gradualy diminished in an upward direction, thus corresponding to the arrangement of the motor centers in the Rolandic area. The accompanying muscular rigidity was extreme. By analogy one might say that such cases corresponded to the spastic type of Little's disease in which cortical hemorrhages and thrombosis of the longitudinal sinus and cerebral veins play an important role.

Sensory disturbances were also occasionally noted by Holmes and Sargent and their sensory investigations are of interest as bearing on the symptomatology of lesions of the parietal lobe. Their summary is as follows:

"The sensory disturbances are especially interesting, as they are almost always those of a pure cortical lesion unaccompanied by any shock effect. The appreciation of pain and temperature is unaffected, and there is no definite diminution of tactile sensibility, but a certain number of light contacts are not recognized; there is, however, no threshold alteration, and the proportion of those missed is not directly related to the intensity of the stimulus. On the other hand, the localization of tactile stimuli, the recognition of the position and of passive movements of the limbs, and of form, shape and size, as well as the discrimination of the compass points, may be seriously disturbed. The slightness of the affection of cutaneous sensibility has been frequently astonishing, as many patients have complained spontaneously of numbness and of having 'no feeling' in their legs." One of their cases was of special interest as indicating the possible course of events in the infantile ataxic group, the motor paralysis clearing up rapidly, with persistence of a high degree of ataxia.

It should be emphasized that these studies were made upon adults in whom cortical function was fully developed in contradistinction to the infantile group in which cortical centers and systems are still immature and in which several years must elapse before accurate sensory examinations are possible. This may in part explain the preservation of common sensation in the infantile group, with loss of the power of coördination.

In all other forms of ataxia, save the cerebellar ataxia, there is a disturbance of the deep sensibility, the degree of which usually determines the severity of the motor disorder. This is true of the peripheral, spinal, thalamic and cerebral ataxia.

Why should the deep sensibility be preserved and yet a severe degree of motor incoördination be present in the ataxic birth palsies? No definite answer can be given to this question at the present time which is not more or less theoretical in nature.

In the first place it is possible that the cortical representation for the perception of deep sensibility covers a larger field than is usually held—as is true according to von Monokow of the cortical motor zone, and thus a considerable area might escape injury. As the lesion occurs in early life there is also ample opportunity for compensatory replacements and substitutions on the part of subcortical centers and neighboring regions of the cerebral cortex. It is also possible that the sensory area for the perception of muscle sensibility is limited to the postcentral convolution and so might escape entirely the ravages of the lesion.

In the course of development of the cortical functions of motility the acquired memories of movements and their complex coördination are registered in a manner similar to the storing of the speech memories. These engrams represent a higher sensory elaboration of movement and would be all-important in the carrying out of precise and definite motor acts. If the region of the parietal cortex subserving this higher function of motility were injured on both sides in early life these memories would be improperly registered or the commissural fibers, through which they exert their influence on motility, would be interrupted with a resulting disharmony of movement. This would produce a cortical or transcortical ataxia, a disorder in the sphere of movement similar to apraxia or agnosia, in which the mechanism for the reception or transmission of higher motor memories would be seriously impaired, the bilaterality of the lesion preventing any compensatory activity on the part of the opposite hemisphere.

As the injury to the cortex takes place at birth, and as these cases cannot be subjected to accurate sensory tests much before the sixth or seventh year, there is ample opportunity for restoration and substitution of function. It is therefore not altogether surprising that the deep sensibility is preserved in conjunction with incoördination of the general motor activities. The cortical centers subserving the function of protopathic sensibility either escape injury or are capable of being replaced by the compensatory activity of adjacent cortical areas.

The higher coördination activities of the muscular system are gradually acquired and are especially retarded and slow of development in this group of cases. That even these are susceptible of gradual development is shown by the clinical course of the cases.

Clinical Comments. The cerebral diataxia bears a certain resemblance to the cerebellar ataxia. The gait, like the cerebellar gait, may be somewhat titubating; there is dysdiadokokinesis and the speech has a dysarthric character not unlike that observed in cases of cerebellar disease. There is, however, no nystagmus, the disturbance of movement is not a dysmetria but a true ataxia, and the ataxia of the extremities is marked and persists in the recumbent posture. In the recumbent posture, with the extremities elevated and the eyes closed, there are ataxic movements, such as are observed in spinal ataxia and unlike the true cerebellar ataxia. Furthermore, the ataxia of the distal portion of the extremities is quite evident. There is also, especially during effort and mental excitement, a tendency to atactiform movements, of a more general character, which resemble or rather suggest a choreiform disturbance. They are not, however, spontaneous, but disappear in the relaxed and recumbent posture and appear only during muscular effort and movement. They are to be regarded as compensatory ataxic manifestations, similar to those occasionally observed in other forms of ataxia.

Another point of interest is the presence of a slight degree of hypotonia. It is well known that lesions of the parietal lobe may produce hypotonicity of the musculature, with diminution of the tendon reflexes. It is therefore not surprising to encounter a certain degree of hypotonicity in conjunction with cortical ataxia from lesions of the parietal lobe. In some of the cases described as cerebellar or atonic-astasic types of diplegia the nature of the ataxia and the associated hypotonia are somewhat suggestive of a parietal localization. This opens up a field for future investigation, and it is my belief that certain symptoms of diplegia which have heretofore been relegated to the cerebellum are really of cortical origin and referable to the higher cerebral centers for the control and coördination of movement.

In the above description I have attempted to outline the chief clinical features of the ataxic type of cerebral birth palsy or cerebral diataxia. It is the sensory equivalent of spastic diplegia, and like this affection shows a similar tendency toward improvement. In a pure form it is, however, comparatively rare.

Some of the cases probably recover or show a very considerable degree of improvement, and are then regarded as of cerebellar origin, an interpretation in which I was originally inclined to participate. After a more detailed study of the motor phenomena, however, I feel convinced that the cortical mechanism is the essential structure involved.

THE ATAXIC DIPLEGIA. This point of view finds a further confirmation in the not infrequent combination of ataxia and spastic paralysis in many cases of cerebral birth palsy, cases in which there are symptoms of generalized spasticity but modified by the association of ataxic symptoms. This constitutes a very grave form of the cerebral birth palsy, examples of which may be found in nearly every institution for the care of chronic diseases. The condition of motor weakness and spasticity being accentuated by the loss of the power of coördination.

Two such cases, aged ten and eleven years respectively, with the typical history of injury at birth, are at the present time under observation in my wards at the Montefiore Home and Hospital. Both cases are confined to the wheel chair and are unable to stand or walk. There is a generalized spastic state which is complicated by a profound general ataxia, any attempt to carry out a precise movement evoking a wild disorder of incoördination. There is no true chorea or athetosis, although the nature of the ataxic disorder suggests a choreiform disturbance.

The atactiform movements do not, however, occur spontaneously, but only on attempts at movement. This I believe is an important factor in differentiating the true chorea and athetosis.

Both patients are fairly intelligent and attend the hospital school. There are no epileptic manifestations. The speech is dysarthric, spastic and very difficult to understand. The sensibility, both superficial and deep, is normal. As in the pure ataxic type there is no demonstrable disorder of the sense of posture or the sense of movement. The stereognostic sense is preserved and any common object placed in the hand is promptly recognized. In these cases I believe there is a combination of lesions which involves the motor sphere causing general rigidity and also the parietal area with a resulting disturbance of coördination. The clinical picture is one of generalized spasticity and ataxia in combination, namely, a *diataxic diplegia*.

Many such cases are, I believe, regarded as choreiform or athetoid types of diplegia, the extreme atactiform disorder of movement simulating very closely chorea. If there is marked spasticity an athetoid tendency is suggested.

In a previous communication I presented the pathological evidence, indicating the relationship of chorea and athetosis to the caudate nucleus and putamen of the corpus striatum, and there can be no question as to the paramount importance of the corpus striatum in the production of true chorea and athetosis. While the relationship of the cerebral cortex to chorea and athetosis is accepted by all systematic writers, the exact localization of this function is still one of the unsolved problems of brain pathology. In all such investigations the existence of a cortical diataxia should be carefully considered and its very close resemblance to such disorders as chorea and athetosis.

CONCLUDING REMARKS. A variety of clinical types of the cerebral palsies of childhood are now recognized, and as such are firmly established in medical literature. Important among these may be mentioned the various forms of the spastic type of Little, the atonic or flaccid type of Förster, and those of cerebellar origin.

In this paper I have directed attention to the existence of a pure ataxic type which may be distinguished from other forms as *diataxia cerebralis infantilis*.

The infantile cerebral diataxia is a special form of the cerebral birth palsy in which the symptomatology is characterized by a generalized disturbance of coördination without evidences of paralysis, spasticity, epilepsy or serious mental defect. The sensibility, both special and general, is apparently unaffected.

The clinical picture is believed to be dependent upon a bilateral vascular lesion in the parietal region, viz., hemorrhage and softening in the distribution of the parietal veins, resulting from an injury at birth.

The parietal area of the cerebral cortex is concerned with the reception and elaboration of higher motor memories, and a bilateral lesion in this region in early life would seriously interfere with the proper development of its cortical centers, association fibers and commissural systems. This would produce a disorder of the higher cortical functions of coördination—with a resulting *cortical* or *transcortical ataxia*.

The disturbance of coördination affects the speech, the gait and station and the use of the extremities, and is a pure ataxic disturbance.

It may be distinguished from cerebellar ataxia by the character of the motor disorder, its persistence in the recumbent posture and the absence of nystagmus. It may be regarded as the sensory equivalent of the spastic type of Little and is not infrequently encountered in combination with this affection, constituting a *diataxic diplegia*.

As in Little's disease, there is a tendency toward improvement, although the development of motility is always seriously retarded.

In its more severe forms this disorder of coördination may be associated with a tendency to atactiform movements which resemble superficially the phenomena of genuine chorea and athetosis; the recognition of which I believe to be of considerable importance in the eventual solution of the problem of the nature and localization of chorea and athetosis.

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ANGINA ABDOMINIS.[1]

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The term angina abdominis was introduced by Baccelli to describe painful seizures in the abdomen due to aneurysmal for-

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[1] Read before the American Gastro-enterological Association.